ASTHENIC BULBAR PARALYSIS.*

By Wharton Sinkler, M.D.

This peculiar disease was first described by Wilks, in 1870, in *Guy's Hospital Reports*[1], where he relates two cases which he speaks of as "an unusual form of glosso-labio-laryngeal paralysis." In 1877[2] he described another case of disease of the medulla, in which death supervened within a few weeks from respiratory paralysis, and a careful autopsy revealed no perceptible lesion. In 1878, Erb[3] published three cases, in which the following symptoms were peculiarly prominent:—bilateral ptosis, isolated in two of the cases, and in the third the intrinsic muscles of the eyeballs were also affected. There was weakness of the muscles of mastication, paresis of the neck muscles, and weakness of the tongue and limbs. The upper branches of the facial nerve were affected, while the lower remained intact. There was no muscular atrophy. The paretic symptoms fluctuated in intensity, improving by rest, and becoming aggravated by fatigue and exertion. Erb called the disease "A new syndrome, probably of bulbar origin." He was evidently not acquainted with the publication of Wilks' papers. Nothing more was added to the subject until 1887, when Oppenheim[4] reported a case with autopsy. Soon after this Goldflam[5] published a paper, in which he collected all previous cases, and added four new ones. To Goldflam belongs the credit of having first called attention to the importance of the fact that the morbid phenomena in this affection are essentially variable in intensity, and are rather due to peculiar weakness of the muscles, evidenced by their quickly giving out as a result of the slightest effort, than to general paralysis. In 1892 Hoppe[6] wrote a paper, giving a full and complete account of the disease, and from this time more attention was drawn to it. Strümpell[7] in 1895 reviewed the literature of the subject, and collected 21 cases which had been reported. Various names have been given to the disease, some of which are "Bulbar Paralysis, without Anatomical Lesions at Autopsy;" "Myasthenie grave pseudo-paralytique" (Jolly); "Syndrome d'Erb et Goldflam;" "Syndrome de Hoppe et Goldflam;" "Maladie d'Erb," "Syndrome bulbaire d'Erb;" "Chronic Progressive Bulbar Paralysis (Oppenheim).

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[1] Wilks, Guy's Hospital Reports, 1870, Vol. XXIII.

[2] Wilks, Guy's Hospital Reports, 1877.

[3] Erb, Archiv für Psychiatrie, Vol. IX, p. 336.

[4] Oppenheim and Siemerling, Berl. klin. Wochenschrift, 1886, Vol. XXIII, p. 791.

[5] Goldflam, Deutsche Zeitschrift für Nervenheilkunde, Vol. IV.

[6] Hoppe, Berliner klinische Wochenschrift, 1892.

Strümpell has called the affection "Asthenic Bulbar Paralysis," and this seems to have found general favor, as it is the name now generally used. In addition to the 21 cases collected by Strümpell, there have been 13 others reported. Of these, but three were by Americans—one by Collins[8], one by Wheaton[9], and one by Berkley[10]. The other cases recorded were by Marie[11], Ballet[12], Cardarelli[13], Finizio[14], Widal and Marinesco[15], Silbermark[16] (2 cases), Brissaud and Lantzenberg[17] (2 cases), and Charcot and Marinesco[18] (1 case).

The principal features of the affection are as follows: The disease appears usually before thirty, and may occur as early as the age of 12 or 15 years, with perhaps a slight preference for the female sex. It begins gradually and without apparent cause, but there may be quite rapid increase in the symptoms, and it may result fatally within a few weeks. Occasionally, there is at first pain, never very violent, and mild vertigo. An early and most important symptom is the rapid fatigue usually shown in those muscles whose nerve centers are in the medulla, pons and crura. The first symptom may be drooping of the lids and diplopia, or else difficulty in mastication, speaking or swallowing. Involvement of the trunk and limbs usually occurs later, although the disease may begin in these parts. There are seldom muscular twitchings observed even in the tongue. When the disease is fully developed, there is permanent paresis in the upper lids and all the muscles innervated by the seventh nerve, and in the muscles of mastication. In the muscles elsewhere there is great and rapid fatigue after the slightest effort, which, in the later stages, amounts almost to complete paralysis. This is noted especially in swallowing, chewing and speaking, and it is also true of the movements of the limbs. The permanent paresis is found in those muscles which are continually in tonic contraction, and have little rest; that is, in the levatores palpebrarum, the muscles of expression, and the masseters. In some cases permanent paralysis of the muscles of the eyeballs is present, and these are also in constant tonicity; often, however, these muscles are entirely spared. Of the muscles below the head, only those of the back of the neck have been found permanently paretic, the explanation for this being, that they are in the same condition of tonic contraction as the muscles of the lower jaw. Ptosis is often the first symptom, and when the muscles of the eyeballs escape, it is important as showing an involvement of one branch only of the third nerve. Reaction of the pupils to light and accommodation always seems to be normal. The extremities are always involved, but weakness of the muscles of the organs of respiration, if it occurs at all, appears late, and the sphincters have never been found affected. On certain days the patient has more power than on others, but anything causing unusual fatigue or depression impairs the strength of the muscles. For example, in the case related by Wheaton, and in my own case, during the menstrual period the paresis was always greater. Irregularity of the heart's action, and tachycardia are sometimes met with. Sudden relapses are common, and may occur even one or two years after apparent recovery.

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[7] Strümpell, Deutsche Zeitschrift für Nervenheilkunde, Vol. VIII, p. 16.

[8] Collins, Joseph, International Medical Magazine, April, 1896, p. 203.

[9] Wheaton, Boston Medical and Surgical Journal, 1898, p. 54.

[10] Berkley, Johns Hopkins Hospital Reports, 1897, p. 94.

[11] Marie, Bull. et mem. soc. méd. des hop. de Par., 1898, 3 S., XV, 437.

[12] Ballet, Gaz. hebd. de méd., Par., 1898.

[13] Cardarelli, Clin. méd., Pisa, 1898, IV, 81.

[14] Finizio, Riforma med., Napoli, 1898, XIV, Pt. 1, 589.

[15] Widal and Marinesco, Bull. et mem. soc. méd. des hop. de Par., 1897, 3 S., XIV, 518.

[16] Silbermark, Wiener klinische Rundschau, 1896, X, 767.

[17] Brissaud and Lantzenberg, Arch. gen. de méd., Par., 1897, I, 257.

[18] Charcot and Marinesco, Compt. rend. de la soc. de biol., March 1, 1895.

About fifteen autopsies have been made, and in none have microscopical changes ever been found in any of the tissues, and from the clinical conditions gross lesions could not be expected. Most writers suggest the theory of infection by toxic materials, although no one can say if the poison is elaborated in the body or taken from without. It is exclusively an affection of the motor system.

The same exhaustion is shown to the faradic current, which proves that the peripheral neuron is involved perhaps alone, although the disease may be muscular. The knee-jerk is prompt, and muscular atrophy does not occur, even after a long period.

The prognosis naturally is doubtful, but the patients usually have periods of remission, and are frequently apparently quite well for several years at a time. There are a number of essential differences between this affection and true degenerative bulbar paralysis. There is no real atrophy of the muscles, either of the tongue, lips or extremities, and there is no complete paralysis of the muscles of the lips producing drooling or drivelling of saliva. The patient is usually able to expose the teeth, pucker the mouth, and to whistle. The deep reflexes are preserved, and there is no change in electrical irritability, except that it becomes exhausted after the continuous application of the current. There are no disturbances of sensibility and the special senses are unaffected. If, however, prolonged stimulation of the special senses is persisted in, they are liable to become quickly exhausted. For example, sight and hearing become impaired after long use. In true bulbar paralysis there is seldom involvement of the third nerve, or the lower facial, or minor branch of the fifth nerve.

Amyosthenia, or weakness of the lower extremities, is not observed in bulbar paralysis, and the attacks of disordered heart's action and dyspnea, which have been recorded as frequently occurring in asthenic bulbar paralysis, are seldom met with in the former disease. As already pointed out, the most characteristic feature of asthenic bulbar paralysis is the readiness with which the affected muscles become exhausted upon use. The patient, whose case I shall record, talks quite distinctly and readily at first, but after a few minutes' conversation her voice becomes weak, high-pitched and nasal. In mastication of food the muscles soon become exhausted, and the patient has difficulty in swallowing the bolus. The movements of the arms and legs also cause feebleness in a short time. The knee-jerks, which are active on first examination, soon become exhausted. Jolly and Murri[19] have pointed out the peculiar influence of electricity upon the muscles. At first, to the faradic current, these respond energetically, but after the passage of the current for a short time, the response becomes very feeble or nil. This Jolly called the "myasthenic reaction."

The opinion of Strümpell and others as to the pathology of the disease is, that it is infectious in character, and that the presence of some poison in the system exerts its deleterious influence upon the groups of ganglionic cells in the medulla and pons. Nothing so far has thrown any light upon the causation of asthenic bulbar paralysis.

It seems to me that a fact in connection with the case which I am about to relate throws some light upon the pathology of the disease. This fact is, that two brothers of my patient had nystagmus and nodding tremor of the head, and two of her own children have been affected in precisely the same way. This would suggest that there was a hereditary predisposition to lesions in the nervous system, and that there existed a developmental defect in the motor tracts of the medulla and pons. We cannot help feeling that, as we study cases of this kind, and observe the history of heredity, how strong is the influence in the direction of hereditary transmission. The number of familiar diseases of the nervous system is increasing each year. and we are beginning to learn that a certain class of diseases is liable to attack individuals in whom there is a poorly constructed nervous system.

The possibility of hysteria being mistaken for this disease is quite great, especially when ptosis and diplopia are the only symptoms, and disappear and reappear without apparent cause. One should always be guarded in making a positive diagnosis of hysteria before carefully considering the possibility of asthenic bulbar paralysis. In the early stages of the disease under consideration, it might readily be mistaken for nuclear ophthalmoplegia, but in asthenic bulbar paralysis the paralysis is not so complete, and after rest the muscles involved always regain a certain amount of power. Moreover, the muscles of speech and deglutition sooner or later become affected in the latter disease.

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[19] Cited by Brissaud and Lantzenberg, Arch. gen. de méd., Par., 1897, Vol. I.

The following case typically exemplifies the description given of asthenic bulbar paralysis by Erb, Strümpell and others:

The patient, Mrs. H. B——, was brought to my clinic at the Orthopedic Hospital and Infirmary for Nervous Diseases by D. C. B. Hough, of Hamburg, Penna. The patient was 37 years of age and married. She was born in America of Scotch parents. Her father died of apoplexy, and her mother is living and well at 65 years. Her maternal grandmother was blind for years before her death. She has two brothers and four sisters who are living, and who are in good health, with the exception of one brother who has nystagmus and a rotary tremor of the head. One brother who died at 30 years of age also had nystagmus and head shaking. She has been married for 16 years. Her eldest child is a boy of 15, who, like the maternal uncles above referred to, has nystagmus and a rotary tremor of the head. He is very small for his age, and undeveloped. He does not look more than 12 years of age, but he is active and very intelligent. His mother says that he does the work of a man on the farm where they live. Her youngest child is five months old, and this one also has nystagmus. The remaining four children are normal. Her husband is living and healthy. There is nothing in the history to give rise to any suspicion of syphilis.

Previous History.—There is nothing notable in the early history of the case. The patient had the ordinary diseases of childhood in mild form. Seven years ago she had a light attack of typhoid fever, and a short time after this, without any exciting cause, she gradually developed ptosis, first in the left and then in the right lid. This remained for at least six weeks. She went to the Wills' Eye Hospital in this city, and under treatment the ptosis gradually recovered. The history taken from the records at the Wills' Eye Hospital states that the lids commenced to droop in September, 1892. On January 30, 1893, when she was admitted to that institution, there was complete ptosis. After six weeks' treatment in which mercury was administered, there was scarcely any drooping of the lids. In October, 1895, there was a similar attack, and her doctor wrote for advice, as she could not come to the hospital. In October, 1897, she returned with diplopia gradually progressing. There was then paresis of both interni, and faintly so of the superior recti muscles. The fields at this time were much contracted. In 1898, except that there was occasional drooping in the lid of the left eye, which is especially marked on near work, nothing seemed wrong. Still apparent paresis of muscles.

A child was born in December, 1898. It was not a difficult labor. She states that, a few days after the birth of her child, her eyesight began to fail more rapidly, and a month later she could not swallow well. Then her hands and arms began to lose power, and she also had feebleness in the legs. She has never been confined to bed. The weakness in her throat muscles, difficulty in swallowing and enunciating have been the most advanced symptoms. She has no pain, but some distress in the lumbar region. She has always had control of the bowels and bladder. She has not nursed her baby, and has menstruated regularly since its birth. At each catamenial period all her symptoms are more marked.

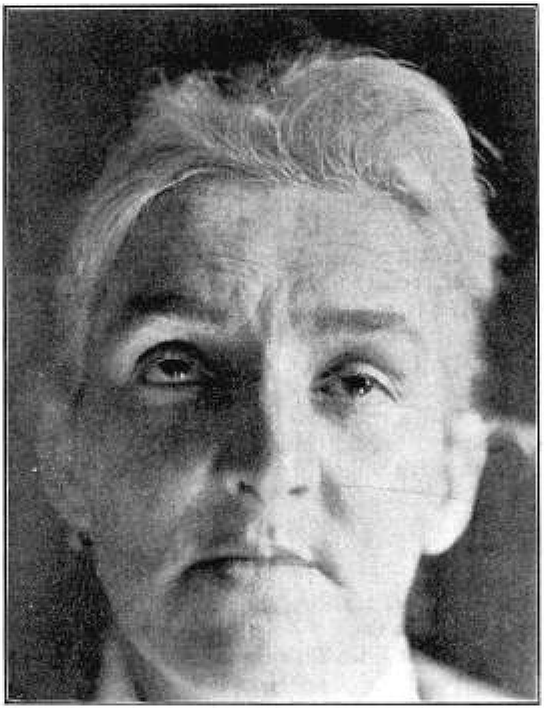

FIG. I. Asthenic bulbar paralysis. Shows paralysis of left superior rectus in looking upward.

On examination, the peculiar expression of the woman's face is noticeable. The eyes are partly closed; the brows being elevated so as to raise the lids to give sufficient vision to go about. The face is without expression and mask-like. Absence of the upper teeth give rise to a peculiar sinking of the upper lip. There is no paralysis of the facial muscles. The tongue is protruded straight. The lips can be separated so as to show the gums, and the mouth can be puckered into the position of whistling. Her speech is nasal, indistinct and whining. After speaking for some time, almost complete aphonia exists. The effort of talking soon exhausts the power of articulation, but after resting for a time she is able to enunciate quite distinctly. Deglutition is difficult. After chewing and swallowing for a short time, the effort of swallowing becomes great,

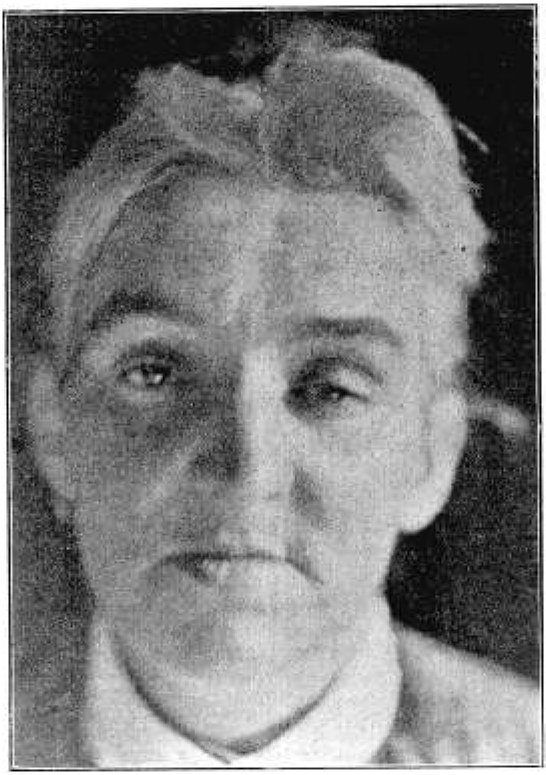

FIG. II. Asthenic bulbar paralysis. Face at rest.

and sometimes liquids are regurgitated through the nose. On inspection the throat muscles appear flaccid. Her hearing is good, and the special senses are unchanged. The arms are weak and soon tire after exertion. She walks in a shuffling and awkward manner, and the strength of the legs soon gives out and the gait becomes more unsteady. The knee-jerks are preserved and apparently do not become exhausted on repeated testing. The sensation is unimpaired, both as to tactile and pain sense. There is no thermo-anesthesia. Electrically, all of the muscles respond to the faradic current, and although not tested extremely, faradic irritability is not exhausted, as was observed by Jolly and others. There is no atrophy of the tongue, nor, in fact, of any of the muscles, and there is no fibrillary tremors in any part. There is no kyphosis of the spine. Dynamometer, right, 50; left, 50. The patient suffers no trouble with her digestion. The bowels are regular. She sleeps well and has no headaches.

Dr. A. G. Thomson made an examination of the eyes and made the following report: "Partial ptosis in both lids; more marked in the left. Pupilary reactions normal. Almost complete paralysis of the left external rectus, and partial paralysis of the left superior rectus. The accommodation is normal. The discs and fundi are normal. Form and color fields are unchanged."

There is apparently no mental disturbance, and the patient is of cheerful and contented disposition in spite of her infirmaties. The fatigue which occurs in the various muscles, both the muscles of the eye and throat, and those of the legs and arms, is markedly characteristic of the affection, and exhibits the distinctiveness of the disease from organic bulbar paralysis.

172. STOMATITIS NEUROTICA CHRONICA (Neurotic Ulcers of the Mouth). By W. Knowsley Sibley (British Medical Journal, No. 1998, page 900, April 15, 1899).

These ulcers have been described by a number of authors under various titles. Their principal claims to be neurotic seem to be that they occur in neuropaths; are aggravated by emotional and mental disturbances; are peculiarly chronic and recurrent; and are not easily explained on any other theory. The present custom seems to be to label the unexplained "neurotic." The author reports three cases, all in women beyond middle age. PATRICK.